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ABSTRACT

This booklet explains characteristics of Marfan Syndrome, an inherited disorder of connective tissue which can be life-threatening if untreated. Medical problems affecting various parts of the body such as the heart, the skeleton, the eyes and the skin associated with Marfan Syndrome are discussed. Possible medical emergencies are identified. Specific learning problems which may affect some children with Marfan syndrome are noted. Other topics considered include participation of children with Marfan syndrome in physical activities, how teachers can help these children, cause of Marfan syndrome, and treatment of Marfan syndrome. (DB)

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THE MARFAN SYNDROME:

A BOOKLET FOR TEACHERS

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THE MARFAN SYNDROME:

A BOOKLET FOR TEACHERS

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Acknowledgement

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INTRODUCTION

The Marfan syndrome is an inherited disorder of connective tissue affecting approximately 40,000 people in the United States. It affects both males and females in equal proportion and is present in all ethnic groups. If untreated, the Marfan syndrome can be a serious life-threatening condition. However, recent advances in treatment have allowed affected individuals to lead long, productive lives. The disorder is not contagious. Since the condition is genetic in origin, it is present from birth in those individuals who are affected.

Many teachers have contacted the National Marfan Foundation with questions about the Marfan syndrome, ranging from concerns about medical aspects of the disorder, to questions about learning disabilities and their possible association with the syndrome. This booklet was prepared to answer some of the questions asked most commonly by teachers. Additional information on medical aspects of this syndrome is available from the National Marfan Foundation.

WHAT MEDICAL PROBLEMS ARE ASSOCIATED WITH THE MARFAN SYNDROME?

Although many medical problems can be associated with the Marfan syndrome, most affected people will not show all the signs and complications of it. The heart, eyes and skeleton are usually involved in most people with the Marfan syndrome, but some will not have symptoms in all three organ systems, or involvement may be mild in one system and more severe in another.

❑ THE HEART

Although the eyes and skeleton are often affected, the most serious problems associated with the Marfan syndrome involve the

heart and blood vessels. A feature of the Marfan syndrome which is present in approximately 60% of affected individuals is "mitral valve prolapse." As a result, blood can leak backwards through the valve of the heart if the leak is severe. People with mitral valve prolapse may have other symptoms such as excessive breathlessness on exercise, a rapid or irregular pulse, or extreme tiredness.

The aorta (the main artery carrying blood away from the heart) is generally wider and more fragile in people with the Marfan syndrome. Aortic dilatation (or widening) is present in approximately 85% of affected people. This widening, which may be present in childhood, is gradually progressive and may ultimately result in the development of tears (dissection) in the wall of the aorta. The gradual widening of the aorta generally occurs without any outward symptoms. As the aorta widens, a tear can develop in its wall. This is quite uncommon in children. However, when this does occur, it is generally associated with severe pain in the front of the chest, in the neck, in the back, or in a combination of sites. When the aorta becomes greatly widened, surgical repair is effective in reducing the risk of catastrophic events. Most patients with the Marfan syndrome will not need cardiac surgery until adulthood.

As the aortic valve stretches over time, blood may begin to leak back into the heart causing it to work harder to pump blood. Without treatment, the heart can become seriously overworked and reach the stage of heart failure. At this stage, the individual may become fatigued with minimal exertion, may faint or develop chest pain.

Because of the increased strain on the aorta produced by vigorous heart action, all Marfan patients should avoid lifting heavy objects, isometric exercise (straining against an immovable resistance), and exercise at maximal capacity. Because the aortic wall is weaker than normal, all contact (collision) sports should be avoided.

❑ THE SKELETON

The skeleton is affected in virtually all individuals with the Marfan syndrome. Most children with the Marfan syndrome are tall for their age, unusually slender and somewhat loose-jointed.

The arms, legs, fingers and toes may be disproportionately long when compared with the trunk. Abnormal spinal curvature (scoliosis) is common and may become quite severe without treatment. The breastbone may often either protrude or indent (pectus deformity).

Children with the Marfan syndrome need to be monitored carefully for curvature of the spine. Abnormal curves tend to worsen during growth, particularly around puberty. Bracing may be effective in stabilizing the spine, but surgery may be necessary if the curvature worsens despite bracing. Some children with the Marfan syndrome have been treated with hormones to advance the age at which puberty begins in order to reduce the number of years during which the spine may deform and to reduce adult height. If such treatment is used, the child must deal with the social and psychological problems of becoming physically and sexually mature before his or her peers.

❑ THE EYES

Most affected children are nearsighted and astigmatic, and untreated, many patients develop one or two lazy eyes (amblyopia). Careful refraction is essential to guarantee vision in both eyes and patching may have to be instituted and vigorously maintained. Impaired muscle balance, especially turning out of a lazy eye is common. The lenses of the eyes of patients with the Marfan syndrome are dislocated in 65% of patients. Surgical removal of the dislocated lenses is not recommended unless they are cataractous (common after age 40 years) or lead to recurrent bouts of acute glaucoma. Holes or tears in the retina occur in approximately 10% of patients and if untreated, resulting retinal detachments lead to blindness. Acute or chronic glaucoma may occur prior to adult life and even prior to age 10 years, requiring medical or surgical treatment.

❑ ADDITIONAL FEATURES

In addition to the features of the Marfan syndrome discussed above, a small percentage of affected individuals will have a spontaneous collapse of a lung (pneumothorax). People with the Marfan syndrome are prone to develop stretch marks in the skin which, although of cosmetic concern, pose no health risk.

WHAT MEDICAL EMERGENCIES MAY ARISE?

While it is unlikely that school personnel will ever need to deal with a medical emergency resulting from complications of the Marfan syndrome in a student, there are some symptoms which need immediate medical attention when they occur:

(1) Severe chest pain, dizziness or unconsciousness - this may indicate aortic problems or irregularities of the heart rhythm.

(2) Severe breathlessness - this may be indicative of a collapsed lung.

(3) Acute eye pain or loss of vision - this may be an indication of pupillary block glaucoma or a retinal detachment.

DO CHILDREN WITH THE MARFAN SYNDROME HAVE LEARNING PROBLEMS?

Marfan syndrome is not associated with mental retardation. However, there is some evidence that children with the syndrome may be at increased risk for learning disability and attention deficit disorder. In addition, there appears to be an increased incidence of verbal-performance discrepancy, with performance scores being lower than verbal scores.

Because of the increased frequency of these kinds of neuropsychological deficits in children with the Marfan syndrome, consideration should be given to screening children for attention and learning problems. Teachers should be made aware of potential difficulties due to hand-wrist hypermobility which are associated with reduced scores on performance tests. In formal school testing situations, children with the Marfan syndrome may need additional time so as not to be inappropriately penalized. Alternate forms of assessing performance requiring less motor input may be

helpful. Children with significant hand-wrist hypermobility may need extra writing time on tests. Wrist or thumb splints to stabilize loose joints may be helpful, and large or padded pens or pencils may allow for a loose grip to be effective.

A child with the Marfan syndrome who is particularly nearsighted may have difficulty seeing the blackboard. When this occurs, teachers should try to distribute a printed copy of the material covered on the blackboard to the student. A larger-than-normal desk may be needed by those students with the Marfan syndrome who are unusually tall.

Because children with the Marfan syndrome are taller than their peers, there is a tendency to treat them as being older than they are, and to have unusually high expectations of them. It is important for teachers to remember their student's chronological age and to treat them appropriately.

CAN CHILDREN WITH THE MARFAN SYNDROME PARTICIPATE IN PHYSICAL ACTIVITIES?

Most young children with the Marfan syndrome, like all children, should participate in physical education. However, there are some activities that should be avoided and an adapted program will need to be worked out. In general, children with the Marfan syndrome should avoid activities that require vigorous exertion, heavy lifting, blows to the head or chest or excessive strain on joints and ligaments. Common sense should prevail when designing sports programs. For example, swimming is not contraindicated, but should be done at an easy pace with opportunity for resting. As much as possible, children with the Marfan syndrome should not be singled out as different and should not have to sit out of gym class away from their classmates.

Unfortunately, two sports in which children with the

Marfan syndrome may excel because of height, basketball and volleyball, are contraindicated because of the amount of contact, the strain on the heart, and the potential for damage to joints and ligaments. Other sports to be avoided are football, boxing, wrestling, weightlifting and hockey. Sports which may be provided through an adapted program include swimming, archery, stationary bicycling, billiards, croquet, darts, golf, bowling, horseshoes and shuffleboard. The affected individual should have medical clearance from his/her cardiologist prior to engaging in sports. In addition, an appropriate adapted physical education program may require input from the orthopedic surgeon and/or ophthalmologist.

HOW CAN TEACHERS HELP STUDENTS WITH THE MARFAN SYNDROME?

Excessive height, feeling different, and inability to participate in some peer activities may lead to a sense of isolation, frustration and depression for the child with the Marfan syndrome. The teachers should recognize that such feelings are common in children who are in any way different from others. Helping a child who feels different from his peers may be addressed by frequent discussion with the child about his or her feelings, and by encouraging the development of other talents and participation in activities which are not restricted. Many children with the Marfan syndrome choose not to discuss their condition and may be embarrassed by having their teacher know about it. However, a teacher's easy acceptance and understanding of the diagnosis will go a long way towards helping the affected child accept it. Without singling out the student with the Marfan syndrome, teachers may want to lead a class discussion about disability and differences in general.

Students with the Marfan syndrome should be guided into a suitable vocation taking into account their special talents. They will need to avoid careers involving heavy exertion or lifting so, for example, a job in construction or as a professional athlete would be inappropriate.

WHAT CAUSES THE MARFAN SYNDROME?

The Marfan syndrome, classified as an autosomal dominant condition, is caused by a change in one gene, present in every cell of the body. The gene causing the Marfan syndrome can be inherited from a parent who also has the condition, or can occur only in the egg or sperm of an unaffected parent (a "new mutation"). About 3/4 of children with the Marfan syndrome have an affected parent, and 1/4 have the condition due to a new mutation. A gene responsible for the Marfan syndrome has just recently been identified, and it is likely that scientists will learn a great deal more about the genetics of the Marfan syndrome in the near future. The Marfan syndrome gene causes the condition by altering the connective tissue in the body, producing the changes in the eye, heart, skeleton and other organs.

A person with the Marfan syndrome has a 50-50 chance of passing on the Marfan gene each time he or she parents a child. The Marfan syndrome can vary widely in severity, even within one family, so a child with the Marfan syndrome can have the condition more or less severely that his or her affected parent.

HOW IS THE MARFAN SYNDROME TREATED?

People with the Marfan syndrome should be treated by physicians familiar with the condition and knowledgeable about

how it affects all body systems. Because several specialists may be involved in the care of each child, absence from school for medical appointments will be essential. There is no cure for this condition yet, but careful medical management can greatly improve prognosis and lengthen life span.

Every person with the Marfan syndrome should have an annual echocardiogram, or in some cases, more frequently to check the size and function of the heart and aorta. Many individuals with the syndrome, including children, will be prescribed a beta-blocking drug such as nadolol, atenolol, or propanolol which will reduce the strength and frequency of the heart's contraction, thus reducing strain on the aorta. These drugs may also produce fatigue, lassitude or depression. If the aorta becomes greatly enlarged or if the mitral valve function is poor, open-heart surgery may be necessary to replace the aortic valve or to repair a leaky mitral valve. Individuals who have had aortic valve replacement may need to take an anticoagulant medication which increases bruising or may limit activities. Because abnormal heart valves are at increased risk for infection during certain dental and surgical procedures, antibiotics are generally prescribed before and after dental visits and selected surgical procedures.

The loose-jointedness associated with the Marfan syndrome may delay walking in infancy or lead to dislocations of the joints, particularly the kneecap. Joint laxity of the wrist and fingers can delay development of fine motor skills, and interfere with learning to draw and write. The joint laxity tends to improve as a child ages, but occupational therapy can be helpful in severe cases.

Affected children need careful monitoring by an ophthalmologist familiar with the Marfan syndrome. Amblyopia (one, or rarely two, lazy eyes) is common and must be treated early. Many children with the Marfan syndrome will need glasses or contact lenses to correct their myopia and astigmatism. In the vast majority of children with the Marfan syndrome, vision is correctable to at least 20/40 in both eyes. Treatment for dislocated lenses is not usually necessary unless the lens malposition leads to acute glaucoma. The eyes should be protected from injury during work and sports. There is no evidence that activities that involve blows to the

head, such as football, boxing and diving worsen the visual prognosis, but they should be avoided because of the risks of cardiac and orthopedic complications. Because many children with the Marfan syndrome have narrowing of the roof of the mouth and a small jaw, the teeth may be crowded. Most dental crowding can be treated by oral surgery and orthodontia.

WHERE CAN I GET ADDITIONAL INFORMATION?

Additional information, including a booklet on the medical aspects of the Marfan syndrome, a booklet for adolescents, and a manual for physical educators, coaches and physicians is available from the National Marfan Foundation. Local chapters of the Foundation, scattered throughout the United States, meet periodically and offer a wide array of social, educational and fundraising activities.

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